

# Wilkie's Syndrome and Left Adnexal Mass: Unusual Presentation of Duodenal Adenocarcinoma

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## ABSTRACT

Duodenal adenocarcinoma (DACa) is a rare malignancy, the presenting symptoms of which are vague and nonspecific. We report the case of a patient presenting with symptoms of subacute small bowel obstruction whose CT scan revealed i) left adnexal mass and ii) compression of 3<sup>rd</sup> portion of duodenum with reduced aortomesenteric angle consistent with Wilkie's syndrome (WS). Laparotomy in addition revealed a distal duodenal stricture, which showed a well differentiated DACa causing subtotal intestinal obstruction. The ovarian mass revealed adenocarcinoma with similar morphology. Immunophenotypic analysis revealed positive expression of CK 20 and CDX 2 and absence of CK 7 staining in the tumours consistent with Primary DACa with ovarian metastasis. We further concluded that the WS resulted from reduced mesenteric fat pad caused by DACa induced cachexia.

The case highlights the elusive nature of duodenal malignancy and emphasises the importance of meticulous small bowel examination during exploration of ovarian masses.

**Keywords:** Duodenal adenocarcinoma, Ovarian metastasis, Wilkie's syndrome

## CASE REPORT

A 58-year-old female presented with complaints of postprandial epigastric pain, vomiting, anorexia and loss of > 10% of weight in 3 months (rapid weight loss). Physical examination revealed poor nourishment, mild abdominal distension and decreased bowel sounds. Her weight was 48kg and height was 1.64m (body mass index = 18 kg/m<sup>2</sup>). Plain X-ray showed a distended stomach and duodenal gas. Abdominal ultrasonogram revealed fluid filled duodenum. On upper GI endoscopy, Wilkie's syndrome (WS) was suspected as the duodenum was dilated till the third part, beyond which the scope could not be negotiated. WS was further confirmed on CT scan which revealed distended stomach and 1<sup>st</sup> and 2<sup>nd</sup> parts of duodenum with compression of the third portion of duodenum, aortomesenteric distance of < 8mm (Normal: 10 to 28 mm), aortomesenteric angle of < 22° (Normal: 25° to 60°) [Table/Fig-1]. A left adnexal contrast enhancing mass was also detected. With a working diagnosis of WS and left adnexal mass, an exploratory laparotomy was done which in addition revealed a stricture in the distal 3<sup>rd</sup> part of duodenum. Duodenal stricture resection and anastomosis with feeding jejunostomy and left adnexectomy was performed. The right ovary was unremarkable therefore right oophorectomy was deferred and only a biopsy was performed. Laboratory investigations revealed: Haemoglobin 9.4 g/dl, normal liver and kidney function tests and CA -125 levels of 7.31 IU/ml (Normal: 0-35 IU/ml).

On macroscopy, the duodenal resection revealed a grey white circumferential ulcero-infiltrative mass, in the distal 3<sup>rd</sup> part of the duodenum, measuring 3.5 cm (inner circumference) x 1 cm (length) x 0.5 cm (depth), at the stricture area causing subtotal (moderate) luminal compromise [Table/Fig-2]. The left ovarian mass was multinodular, apparently encapsulated and measured 6 cm x 4 cm x 4 cm with predominantly solid, lobulated, yellow grey tan cut surface exhibiting focal minute mucoid cystic areas.

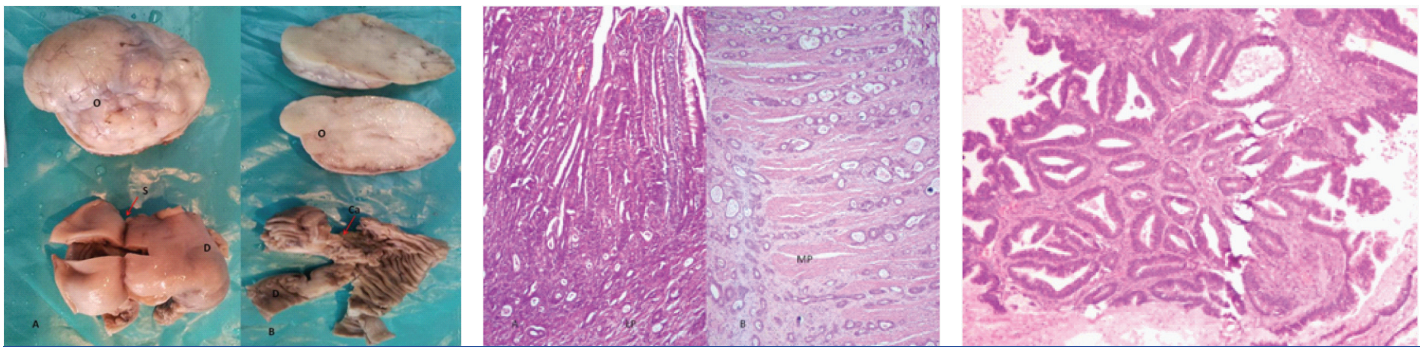
Microscopic examination of the sections from duodenal mass revealed transmurally infiltrating well differentiated duodenal adenocarcinoma (DACa) [Table/Fig-3] and sections from left ovarian mass revealed adenocarcinoma microscopically resembling

the former [Table/Fig-4]. The sections from the right ovarian biopsy were unremarkable. Immunohistochemical staining, done to elucidate the origin and character of the tumour cells revealed positive expression of CK 20 and CDX2 and absence of staining for CK 7 in the duodenal and ovarian tumours [Table/Fig-5]. These histological and immunohistochemical results confirmed the diagnosis of "Primary DACa with left ovarian metastasis; p TNM: T4,N0,M1, Grade I, Stage IV".

The postoperative course was uneventful, and the patient was subsequently referred to oncology for consultation. However she did not follow up as scheduled, and has been lost to follow up care.



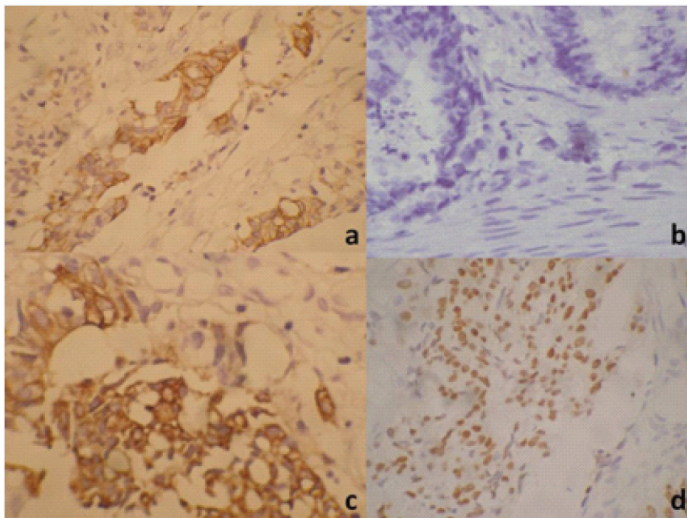
**[Table/Fig-1]:** Contrast enhanced CT scan showing compression of the 3<sup>rd</sup> portion of duodenum (red arrow) between the superior mesenteric artery and aorta with reduced aortomesenteric distance



**[Table/Fig-2]:** (A) External surface of the duodenal resection (D) showing stricture (S) and external surface of the multinodular, apparently encapsulated ovarian mass (O). (B) Cutsurface of the duodenal resection (D) showing grey white circumferential ulceroinfiltrative mass (Ca) and cut surface of the Ovarian mass (O)

**[Table/Fig-3]:** (A) Duodenal mucosa with tumour glands infiltrating the lamina propria (LP). (B) Duodenal muscularis propria (MP) infiltrated by tumour glands. (H and E, x 100)

**[Table/Fig-4]:** Ovarian metastatic adenocarcinoma. (H and E, x 100)



**[Table/Fig-5]:** Duodenal adenocarcinoma displaying membrane and cytoplasmic immunoreactivity for CK 20 (a) and negative staining for CK 7 in the tumour cells (b). Ovarian tumour showing positive membrane and cytoplasmic expression of CK 20 (c) and nuclear expression of CDX 2 (d) in the tumour cells (x400)

## DISCUSSION

Malignant tumours of small intestine are very rare accounting for 0.1 to 0.3% of all malignancies and 1% of all gastrointestinal malignancies [1]. Adenocarcinomas (ACa) account for 36.9% of all small bowel malignancy with the remaining comprised of carcinoid tumours (37.45%), lymphoma (17.3%) and stromal tumours (8.4%) [2].

Duodenal adenocarcinoma (DACa), accounts for < 0.4% of all GIT tumours and 56% of all duodenal malignancies [2,3]. Due to their vague and nonspecific presenting symptoms, a delayed diagnosis or misdiagnosis is common. In a study which included 113 patients with duodenal ACa, the most common clinical presentation was abdominal pain (65%), obstruction (34%) and bleeding (22%) [4]. Other presentations included anorexia, weight loss, abdominal distension, jaundice and diarrhoea [3,5].

In our case the patient presented with symptoms of subacute small bowel obstruction, the cause of which was initially discerned as Wilkie's syndrome (WS) on contrast enhanced computed tomography (CT). WS is an uncommon syndrome, with an incidence of 0.1 to 0.3%, characterised by compression of the 3<sup>rd</sup> portion of duodenum between the superior mesenteric artery and aorta resulting in recurrent mechanical duodenal obstruction [6-8]. Any condition that narrows the aortomesenteric angle can precipitate entrapment of 3<sup>rd</sup> portion of duodenum resulting in WS, which is also known as arteriomesenteric duodenal compression, chronic

duodenal ileus, body cast syndrome or superior mesenteric artery syndrome.

Among the other predisposing conditions like anorexia nervosa, severe injury such as head trauma and postoperative states leading to prolonged bed rest, burns, malabsorption, constitutional factors like thin body built, exaggerated lumbar lordosis, rapid linear growth without compensatory weight gain, abdominal wall laxity and viceroptosis, spinal deformity or trauma, surgical treatment of vertebral fractures or scoliosis entailing use of body casts and anatomic anomalies like abnormally high and fixed ligament of Treitz and low origin of superior mesenteric artery, depletion of mesenteric fat caused by rapid severe weight loss due to catabolic states like cancer is a recognised cause of WS [6,9]. Confirmation of WS requires radiographic procedures such as upper gastrointestinal series, hypotonic duodenography and contrast CT scanning. CT scanning is very useful as, with three-dimensional reconstruction, additionally aortomesenteric angle and aortomesenteric distance can be measured and intra-abdominal and retroperitoneal fat can be assessed. The CT criteria for the diagnosis of WS are aortomesenteric angle of < 22° and aortomesenteric distance of < 8mm which were evident in the current case [6,7,9]. Successful treatment of WS requires identification and removal or reversal of the precipitating cause. Initial conservative management, comprised of adequate nutrition and nasogastric decompression, is recommended. Following failure of the above treatment, surgical procedures like duodenojejunostomy, gastrojejunostomy or division of Ligament of Treitz may be required.

In the present case in addition to causing subtotal obstruction, the DACa resulted in cachexia with consequent reduction in mesenteric fat pad culminating in WS, which further exaggerated the symptoms of upper GIT obstruction. Most of the previous, albeit rare, reported cases of cancer associated WS occurred as complications after operation or radiotherapy [9]. To the best of our knowledge, this is the second reported case of WS caused by DACa [8]. In this report, apart from direct mechanical intestinal obstruction by the tumour, we have presented another mechanism of intestinal obstruction in DACa.

The liver is the most common site of metastasis from small bowel carcinomas and metastasis to ovaries is rare. In studies conducted on 168 patients with ovarian metastasis only four originated from the small intestine [10]. Majority of ovarian metastasis from small bowel primary are misdiagnosed as primary ovarian malignancy and the intestinal primary is diagnosed months after oophorectomy. Thus, meticulous small bowel examination is warranted in patients with ovarian mass and upper abdominal complaints.

The coordinate expression of CK7 and CK20 is useful in differentiating ovarian from intestinal adenocarcinomas. The ovarian carcinomas are CK7 + / CK20 – and the intestinal carcinomas are CK7– / CK20 +. CDX2 is a sensitive marker for intestinal carcinoma which metastasizes to the ovary.

Upper GIT endoscopy, CT scans and barium studies are commonly used to diagnose abdominal diseases. However, these have low sensitivity in diagnosing small bowel tumours. Enteroclysis, capsule endoscopy and double balloon endoscopy are the newer diagnostic modalities which are more sensitive and useful in diagnosing small bowel lesion [1,3].

## CONCLUSION

We have reported a rare case of WS resulting from DACa induced cachexia. DACa should be considered as an etiological factor that may precipitate WS. A high index of suspicion can lead to early diagnosis of these tumours, resulting in timely curative resection which is crucial to an acceptable outcome in these patients. Awareness of the possible precipitating factors of WS and their prompt identification is important as approach for treatment differs according to the aetiology.

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